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De novo adamantinomatous craniopharyngioma presenting anew in an elderly patient with previous normal CT and MRI studies: A case report and implications on pathogenesis

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ABSTRACT

Adamantinomatous craniopharyngiomas are histologically benign epithelial tumors which arise from embryonic remnants of the craniopharyngeal duct and Rathke's pouch. They are thought to have a congenital origin and are histologically unique from papillary craniopharyngioma. We describe the case of an elderly male who presented with symptoms related to a large craniopharyngioma with previously normal brain magnetic resonance and computed tomography imaging studies. These findings dispute the embryogenic theory that craniopharyngiomas observed in adults develop from the persistent slow growth of embryonic remnants.

1. Introduction

The pathogenesis of craniopharyngiomas was first described by Erdheim in 1904 as a tumor developing from squamous epithelial cell rests from a partially involuted craniopharyngeal duct [1]. Classically the incidence of these tumors occurs in a bimodal distribution corresponding to two distinct subtypes: papillary, which is indolent and is most often seen in middle aged patients, and adamantinomatous, which is thought to be more aggressive, typically manifesting in childhood or adolescence [3]. Both subtypes of craniopharyngiomas have been postulated to share a congenital origin resulting from epithelial cells deposited during fetal life along the course of the involuting hypophyseal duct. (ref. [5–7], 9 Argiinteanu) We describe a rare case of a de novo adamantinomatous craniophayrngioma appearing in an elderly patient who had normal cranial MRI and CT imaging studies several years prior to the development of his tumor. This challenges the belief that these tumors arise in adults after slow, continuous growth which begins in childhood.

2. Case history

The patient is a 66-year-old male who presented with decreasing visual acuity over several months. He was referred for a brain MRI which demonstrated a suprasellar mass with a small heterogeneously enhancing solid component, vascular encasement, and a larger cystic component measuring approximately 3.5 cm × 3.6 cm × 3.1 cm most consistent with a craniopharyngioma. This mass was not seen on a head CT obtained 3 years prior as part of a trauma evaluation and MRI completed 7 years prior for evaluation of possible stroke. (Fig. 1) These studies were all obtained at the same center. Endocrinological workup was normal at the time of presentation.

3. Operative and pathological findings

The patient underwent a standard left pterional craniotomy. Gently retracting the left frontal lobe we immediately identified large cystic portions of the tumor in the prechiasmatic area as well as over the carotid artery and extending into the proximal sylvian fissure. After deflating and debulking the cystic portions of the tumor the capsule was carefully dissected off of the optic apparatus and carotid artery and was removed. There was a solid component of the tumor under the left optic nerve and under the chiasm extending to the right side that was meticulously removed as well. Gross total resection was confirmed with postoperative MRI scan.

On gross examination the cystic fluid had a classic machine oil appearance. On histological examination there were classic findings of an adamantinomatous craniopharyngioma including a basal layer of small basophilic cells, a stellate reticulum, areas of focal calcification,

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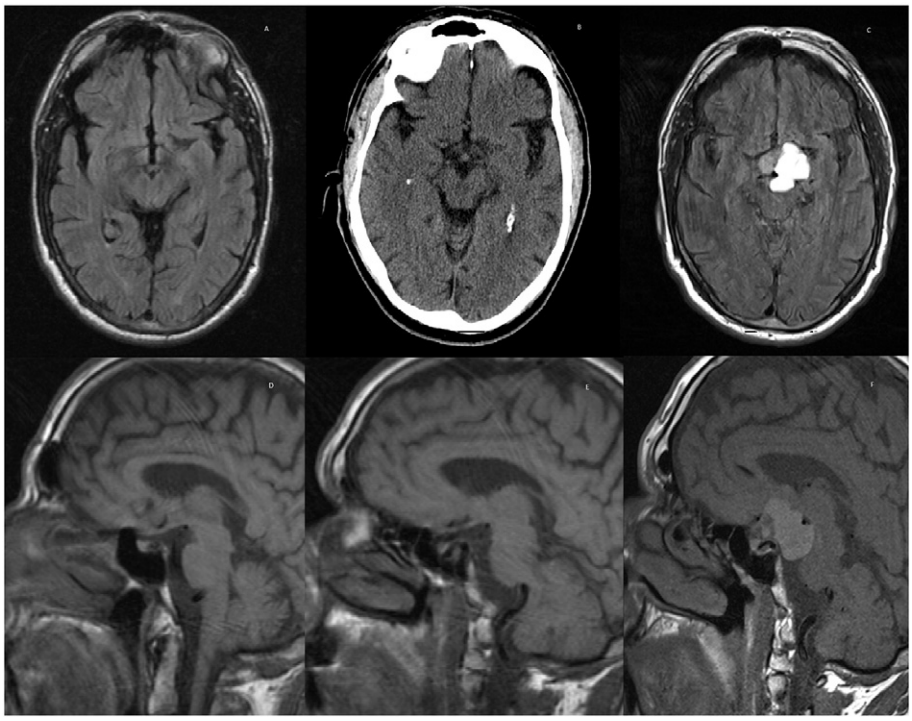

Fig. 1. Axial unenhanced FLAIR MRI 7 years prior (A) and axial unenhanced CT of the brain 3 years prior to diagnosis (B). Axial FLAIR MRI showing the lesion at the time of diagnosis (C).

and extensive deposition of wet keratin. The mass also had strong staining for beta-catenin seen in the adamantinomatous subtype (Fig. 2).

4. Discussion

Classically, craniopharyngioma has been described as a benign neoplasm that develops from cells of a partially involuted craniopharyngeal duct [1]. Zada et al. describe two main subtypes, adamantinomatous and papillary, as lying on a histopathological continuum, with adamantinomatous marking the most aggressive end of the continuum and papillary, the most indolent [6]. Larkin and Ansorge postulate that adamantinomatous craniopharyngiomas are similar to odontogenic tumors whereas papillary tumors may represent a tumor of mature pituitary epithelium, suggesting that these tumors may develop through different pathways.

Two theories have been proposed to describe the etiology of adamantinomatous and papillary subtypes of adamantinomatous craniopharyngioma. The metaplastic theory postulates that differentiated squamous epithelial cells of the adenohypophysis or infundibulum undergo metaplasia resulting in the indolent, papillary craniopharyngioma seen in adults. The embryogenic theory postulates ectopic embryonic remnants of the craniopharyngeal duct undergo neoplastic transformation, which leads to the formation of an adamantinomatous craniopharyngioma in younger patients, and may also account for histological similarities between adamantinomatous craniopharyngiomas and odontogenic tumors [2,3,5,6]. The theory of two separate pathways is supported by mutations in the beta-catenin gene that have been identified in approximately 70% of adamantinomatous but not papillary craniopharyngiomas as seen in our patient. Moreover, 90-95% of adamantinomatous craniopharyngiomas stained positively for nuclear or cytosolic B-catenin, which is absent in papillary craniopharyngiomas. However, other such mutations have not been determined [5].

To our knowledge, only one other case of a de novo adamantinomatous craniopharyngioma has been previously reported [4]. This case challenges the notion that adamantinomatous craniopharyngiomas develop from embryonic remnants that undergo early neoplastic transformation

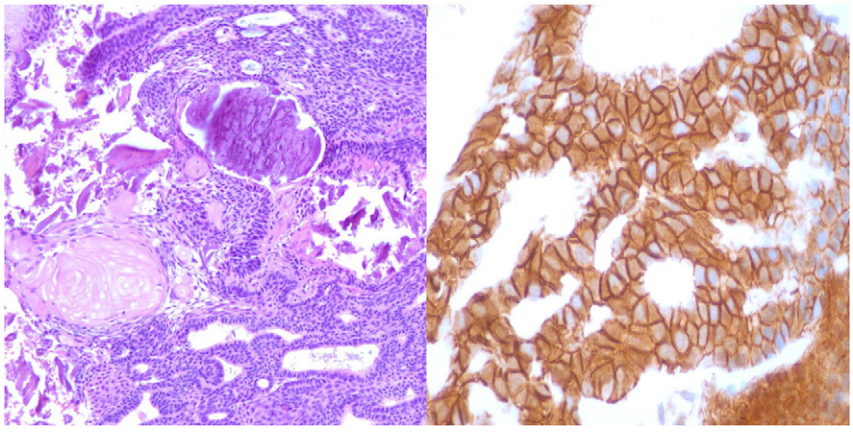

Fig. 2. Surgical specimen showing small basophilic cells, stellate reticulum, areas of focal calcification, and extensive deposition of wet keratin. Hematoxylin and eosin 10× (left). Strong beta-catenin staining in the same specimen. Beta-catenin 60× (right).

causing symptoms and identification of these tumors early in life. If the embryogenic theory is correct, then this patient is likely to have already had ectopic embryonal cells of the craniopharyngeal duct in the suprasellar region throughout his life, and there should be evidence of this on his original imaging. The absence of such findings raises questions as to the natural history of these tumors. One possibility is that a "second-hit" gene mutation caused the sudden growth of this tumor. Although the beta-catenin mutation is found in approximately 70% of adamantinomatous craniopharyngiomas, the other tumorigenic mutations responsible are still unknown. These genes may be responsible for de novo tumors of this subtype for which there is only one other report in the literature.

There is inconclusive evidence regarding the etiology and development of craniopharyngiomas. This report challenges some existing theories and provides evidence that further investigation into genetic, molecular, and embryological pathways responsible for these tumors and their progression is essential.

Conflicts of interest

None. We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome. We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship but are not listed. We further confirm that the order of authors listed in the manuscript has been approved by all of us. We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property. We further confirm that any aspect of the work covered in this manuscript that has involved either experimental animals or human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript. We understand that the Corresponding Author is the sole contact for the Editorial process (including Editorial Manager and direct communications with the office). He/she is responsible for communicating with the other authors about progress, submissions of revisions and final approval of proofs. We confirm that we have provided a current, correct email address which is accessible by the Corresponding Author.

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